A serpentine and hypoplastic infrarenal aorta associated with aneurysm: A case report

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Few congenital anomalies of the abdominal aorta have been well described in the literature. Coarctation of the abdominal portion of the aorta is a relatively rare occurrence that accounts for less than 2% of all aortic coarctations.[1] Although some studies use the term *hypoplasia* interchangeably with coarctation, a distinct hypoplastic aortic syndrome also has been described. These patients are generally women who smoke, who are aged 30 to 45 years, and who have small distal aortas in conjunction with atherosclerotic disease.[2] Hypoplasia of the aorta usually is associated with occlusive disease but not commonly with other aortic abnormalities. In this report, we describe the case of a 62-year-old man with an unusual aortic anomaly that we believe to be congenital in origin: a serpentine and hypoplastic infrarenal aorta found in conjunction with an abdominal aortic aneurysm.

CASE REPORT

A 62-year-old man with an infrarenal abdominal aortic aneurysm was referred to our facility after an interval increase in size of the aneurysm to 5 cm. His medical history was remarkable for poorly controlled hypertension. He had no significant surgical history. Medications included amlodipine besylate, atenolol, and buspirone hydrochloride. The patient had a 60 pack-year smoking history.

A physical examination showed a healthy-appearing older man. Blood pressure was 170/72 mm Hg, and vital signs were otherwise normal. Physical examination findings were remarkable for a pulsatile mass in the abdomen and for lower-extremity pulses that were palpable and symmetric. The patient had ankle-brachial indexes of 1.01 on the right and 0.91 on the left. Results of laboratory studies were unremarkable. Chest x-ray films were normal except for calcifications noted within the thoracic aorta. Abdominal computed-tomography findings included an extremely narrow infrarenal abdominal aorta with a diameter of 1.2 cm at the level of the renal veins. No evidence was seen of atherosclerotic calcification, thickened aortic wall, or periaortic soft tissue mass in this region. We noted a fusiform abdominal aortic aneurysm with a diameter of 5 cm that began 6 cm below the origin of the renal arteries. The aortic bifurcation measured distally 2.7 cm, and both common iliac arteries measured 1.1 cm at their origins. The right common iliac artery had a focal dilation to 1.8 cm but returned to normal caliber. Both common iliac arteries measured 1.1 cm at their bifurcations. The internal and the external iliac arteries had evidence of mild atherosclerotic changes, but both were without narrowing or aneurysmal dilatation. An abdominal aortogram showed a serpentine and hypoplastic lumen of the abdominal aorta distal to the renal arteries and proximal to the aneurysmal segment. An accessory left renal artery was noted (Figs. 1-3).

Preoperative cardiac evaluation indicated significant coronary artery disease with reversible ischemia. Therefore the patient first underwent two-vessel coronary artery bypass grafting. After 8 weeks, the patient returned for elective repair of his abdominal aortic aneurysm.

At the time of operation, the abdominal aorta was exposed through a midline incision. The aorta above the celiac trunk to the origin of the renal arteries appeared grossly normal. From the renal arteries to the neck of the aneurysm, a distance of 6 cm, the aortic diameter was 1 to 1.5 cm. This hypoplastic segment made four distinct curves before the aneurysmal segment. No evidence of periaortic inflammation or fibrosis was found. The small accessory left renal artery was noted inferior to the main left renal artery, and an additional aberrant arterial branch was noted on the right side of the aorta just proximal to the aneurysm. Both of these vessels were ligated. The left renal vein crossed anterior to the aorta at the level of the renal arteries just proximal to the hypoplastic segment. Suprarenal aortic control was obtained because of the abrupt decrease in size of this vessel at the level of the renal arteries. Distal control was obtained bilaterally at the iliac bifurcations because of mild aneurysmal changes of the right common iliac artery. Next, the hypoplastic segment and the aneurysm were opened in continuity in a longitudinal fashion. The wall of the hypoplastic aorta was grossly normal both visually and by palpation. No evi-

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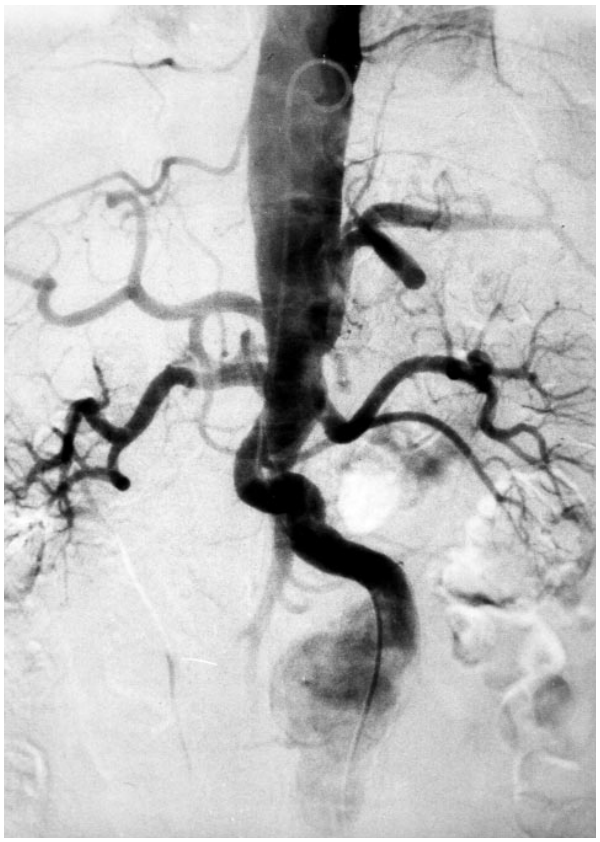

Fig. 1. Aortogram. Anteroposterior view showing patient's serpentine and hypoplastic aorta.

dence was found of inflammation, mural thrombus, or atherosclerotic disease. A single large posterior branch was noted within the hypoplastic segment, which was oversewn. The aorta was transected in a beveled fashion from 1 cm below the renal arteries anteriorly to 0.5 cm above the renal arteries posteriorly. This cuff was anastomosed to a beveled 14 × 7 cm bifurcated double velour-knitted vascular graft, which thereby completely removed the hypoplastic segment. The graft was anastomosed distally to the right and left common iliac arteries just proximal to the bifurcation in an end-to-end fashion. Histologic examination was not performed.

The patient's postoperative course was uneventful, and he was discharged on the seventh postoperative day. He has required the same medication regimen as he had before for control of his hypertension.

DISCUSSION

Congenital abnormalities of the abdominal aorta are unusual. Our literature search yielded accounts of coarctation, hypoplasia, and rare case reports that depicted anatomic variations of aortic branches. The patient we have described had a serpentine and hypoplastic aorta—a distinct entity from the other anomalies that were presented in the literature and one that we believe is congenital in origin.

Coarctation of the aorta is a localized malformation that causes a narrowing of the vessel lumen that is usually severe. Ninety-eight percent of coarctations occur in the descending aorta near the ligamentum arteriosum. Coarctation of the abdominal aorta is quite rare, with an overall incidence ratio of 1:62,500 in an autopsy study, and it comprises less than 2% of all aortic coarctations.[1] Quain[3] is credited with the earliest recognition of this entity; in 1847 he described a stricture of the aorta below the diaphragm that he believed to be congenital in origin. More than 100 years later, in 1952, Glenn et al.[4] reported the first successful surgical correction of this kind of anomaly.

Longer-segment stenoses of the abdominal aorta have been referred to as coarctations or hypoplasia in the literature, and these stenoses can result from either congenital or acquired causes. Maycock[5] suggested in 1937 that congenital long-segment stenoses resulted from unequal fusion of the primitive dorsal aortae. In 1973 Arnot and Louw[6] presented a convincing anatomical argument that supported this theory. These authors studied the abdominal aorta in 100 cadavers, examined the origins of lumbar arteries, and measured aortic diameter at four levels. They found that eight of the cadavers had just one distal bifurcating lumbar artery rather than the usual paired arteries. Each of these same eight cadavers had a significantly more narrow aorta between the origin of the inferior mesenteric artery and the bifurcation of the aorta. The authors concluded that this abnormality resulted from "overfusion" of the two dorsal arteries within the first month of intrauterine development. The finding of multiple renal arteries in many of these patients may support this theory because the formation of a single renal artery occurs during the same time frame and in the same location as the fusion of the dorsal aortae.

The congenitally narrowed distal aorta also plays a role in hypoplastic aortic syndrome, which is described in young female smokers, aged 30 to 45 years.[2] Normally occurring atherosclerosis in combination with a small distal aorta is thought to lead to symptoms at an earlier age. Palmaz et al.[7] presented a retrospective review to study the significance of this disease in men, and they found a good correlation between less-than-normal aortic diameter and early atherosclerosis. They found an association of severe atherosclerotic lesions in the distal aorta with a mean

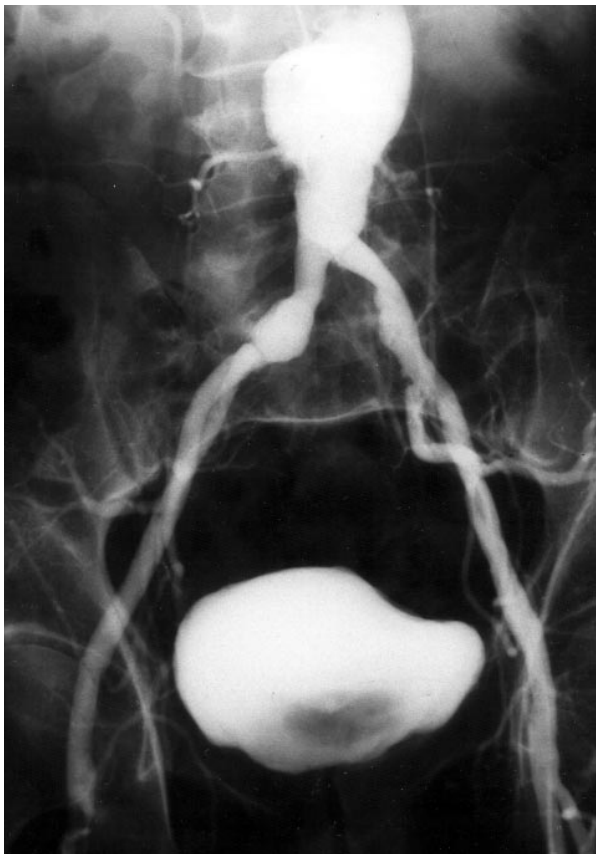

Fig. 2. Aortogram. Anteroposterior view showing aortic bifurcation and common iliac arteries.

Fig. 3. Aortogram. Lateral view. Note the proximal aspect of a single large posterior branch from the hypoplastic segment.

age for onset of symptoms 10 years earlier and a relative absence of generalized atherosclerotic disease. Their findings strongly suggest a mechanical cause for the aorto-iliac lesions in patients with small aortas.

Acquired causes of a hypoplastic segment of the abdominal aorta include forms of nonspecific aortitis, subtypes of Takayasu's arteritis, and radiation aortitis. As with coarctation, these patients also may be first seen with hypertension if the renal arteries are involved, but examination of the involved segment shows marked inflammatory changes. Stenoses related to neurofibromatosis and fibromuscular hyperplasia also have been described, again with the presence of gross and microscopic abnormalities.

The patient in this case report most likely had congenital hypoplasia of his distal aorta. He had an isolated long segment narrowing with a large midline posterior branch arising from this area, possibly representing a common origin of multiple lumbar branches (Fig. 3). Multiple renal arteries can be found in up to 20% of the normal population; however, his accessory left renal artery in association with this hypoplastic aortic segment may well represent an early developmental abnormality during the first month, when dorsal aortae fusion and renal artery formation occurs. Although the serpentine nature of the aorta and the narrowed segment proximal to an aneurysm undoubtedly created wall stress and turbulent flow, the patient surprisingly lacked significant atherosclerotic disease in this hypoplastic area. The aortic wall was soft and of normal thickness; we saw no evidence of atheroma or calcification on examination of the lumen.

We also reviewed the literature that described aneurysms in conjunction with aortic narrowing because his infrarenal aneurysm arose from this area of hypoplastic aorta. Most of the references involved aneurysms in association with coarctation of the thoracic aorta.

Although flow patterns and pressure abnormalities are similar with abdominal coarctations, associated aneurysm formation in these patients has not been

a well-described phenomenon in the literature. Our review found a total of five cases of aneurysm in association with abdominal aortic coarctation, and one of these five patients had multiple aneurysms. In 1972 Sproul and Pinto[8] treated a patient who had a hypoplastic distal aorta and an associated 5-cm saccular aneurysm that arose at the bifurcation. This patient underwent placement of an aortoiliac bypass graft with good results. Pierce et al.[9] described a patient with abdominal aortic coarctation in association with four aneurysms adjacent to or in the hypoplastic segment. The patient was treated with placement of a thoracoabdominal aortic bypass graft and resection of the aneurysms. Most recently, Magnoni et al.[10] reported 20 patients who had abdominal aortic hypoplasia out of a series of 1005 patients who were undergoing surgical treatment for chronic aortoiliac occlusive disease. Three of the 20 patients had aneurysmal dilatation of a hypoplastic aorta.

Aneurysmal degeneration associated with aortic hypoplasia is rare but must be considered when evaluating patients with coarctation. This abnormality generally can be treated by standard aneurysm replacement with synthetic grafts. Although coarctation is not a common cause of hypertension in adults, coarctation of the abdominal aorta must be considered in patients of any age with hypertension. Refractory hypertension, abdominal bruit, and diminished bilateral lower-extremity pulses can provide clues to the diagnosis in cases of more severe disease. Patients with hypoplastic abdominal aortas are more likely to be seen later in life with lower-extremity claudication and weak or absent distal pulses. Surgical correction of both anomalies with aorto-iliac or aorto-bifemoral bypass grafting has achieved good results.

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